

ON A FORM  
OF  
CHRONIC INFLAMMATION OF BONES  
(OSTEITIS DEFORMANS).

BY  
SIR JAMES PAGET, BART., D.C.L., LL.D., F.R.S.,  
CONSULTING SURGEON TO ST. BARTHOLOMEW'S HOSPITAL, ETC.

---

Received November 1st—Read November 14th, 1876.

---

I HOPE it will be agreeable to the Society if I make known some of the results of a study of a rare disease of bones.

The patient on whom I was able to study it was a gentleman of good family, whose parents and grandparents lived to old age with apparently sound health, and among whose relatives no disease was known to have prevailed. Especially, gout and rheumatism, I was told, were not known among them; but one of his sisters died with chronic cancer of the breast.

Till 1854, when he was forty-six years old, the patient had no sign of disease, either general or local. He was a tall, thin, well-formed man, father of healthy children, very active in both mind and body. He lived very temperately, could digest, as he said, anything, and slept always soundly.

At forty-six, from no assigned cause, unless it were that he lived in a rather cold and damp place in the North of England, he began to be subject to aching pains

in his thighs and legs. They were felt chiefly after active exercise, but were never severe; yet the limbs became less agile or, as he called them, "less serviceable," and after about a year he noticed that his left shin was misshapen. His general health was, however, quite unaffected.

I first saw this gentleman in 1856, when these things had been observed for about two years. Except that he was very grey and looked rather old for his age, he might have been considered as in perfect health. He walked with full strength and power, but somewhat stiffly. His left tibia, especially in its lower half, was broad, and felt nodular and uneven, as if not only itself but its periosteum and the integuments over it were thickened. In a much less degree similar changes could be felt in the lower half of the left femur. This limb was occasionally but never severely painful, and there was no tenderness on pressure. Every function appeared well discharged, except that the urine showed rather frequent deposits of lithates. Regarding the case as one of chronic periostitis, I advised iodide of potassium and *Liquor Potassæ*; but they did no good.

Three years later I saw the patient with Mr. Stanley. He was in the same good general health, but the left tibia had become larger, and had a well-marked anterior curve, as if lengthened while its ends were held in place by their attachments to the unchanged fibula. The left femur also was now distinctly enlarged, and felt tuberos at the junction of its upper and middle thirds, and was arched forwards and outwards, so that he could not bring the left knee into contact with the right. There was also some appearance of widening of the left side of the pelvis, the nates on this side being flattened and lowered, and the great trochanter projecting nearly half an inch further from the middle line. The left limb was about a quarter of an inch shorter than the right. The patient believed that the right side of his skull was enlarged, for his hats had become too tight; but the change was not clearly visible.

Notwithstanding these progressive changes, the patient

suffered very little ; he had lived actively, walking, riding, and engaging in all the usual pursuits of a country gentleman, and, except that his limb was clumsy, he might have been indifferent to it. He had taken various medicines, but none had done any good, and iodine, in whatever form, had always done harm.

In the next seventeen years of his life I rarely saw him, but the story of his disease, of which I often heard, may be briefly told and with few dates, for its progress was nearly uniform and very slow. The left femur and tibia became larger, heavier, and somewhat more curved. Very slowly those of the right limb followed the same course, till they gained very nearly the same size and shape. The limbs thus became nearly symmetrical in their deformity, the curving of the left being only a little more outward than that of the right. At the same time, or later, the knees became gradually bent, and, as if by rigidity of their fibrous tissues, lost much of their natural range and movement.

The skull became gradually larger, so that nearly every year, for many years, his hat, and the helmet that he wore as a member of a Yeomanry Corps needed to be enlarged. In 1844 he wore a shako measuring twenty-two and a half inches inside ; in 1876 his hat measured twenty-seven and a quarter inches inside (Pl. I, fig 4). In its enlargement, however, the head retained its natural shape and, to the last, looked intellectual, though with some exaggeration.

The changes of shape and size in both the limbs and the head were arrested, or increased only imperceptibly, in the last three or four years of life.

The spine very slowly became curved and almost rigid. The whole of the cervical vertebræ and the upper dorsal formed a strong posterior, not angular, curve ; and an anterior curve, of similar shape, was formed by the lower dorsal and lumbar vertebræ. The length of the spine thus seemed lessened, and from a height of six feet one inch he sank to about five feet nine inches. At the same

time the chest became contracted, narrow, flattened laterally, deep from before backwards, and the movements of the ribs and of the spine were lessened. There was no complete rigidity, as if by union of bones, but all the movements were very restrained, as if by shortening and rigidity of the fibrous connections of the vertebræ and ribs.

The shape and habitual posture of the patient were thus made strange and peculiar. His head was advanced and lowered, so that the neck was very short, and the chin, when he held his head at ease, was more than an inch lower than the top of the sternum.

The short narrow chest suddenly widened into a much shorter and broad abdomen, and the pelvis was wide and low. The arms appeared unnaturally long, and, though the shoulders were very high, the hands hung low down by the thighs and in front of them. Altogether, the attitude in standing looked simian, strangely in contrast with the large head and handsome features.\*

All the changes of shape and attitude are well shown in sketches from photographs taken six months before death (see Pl. I, figs. 1 to 3). Only the lowering of the necks of the femora is not shown. In measurement after death the axes of the shaft and neck of the right femur formed an angle of only  $100^{\circ}$  instead of  $120^{\circ}$  or  $125^{\circ}$ , and this change of shape added to the appearance of increased width of the pelvis.

But with all these changes in shape and mobility of the head, spine, and lower limbs, the upper limbs remained perfect, and there was no disturbance of the general health.

In 1870, when the disease had existed sixteen years, the left knee-joint was, for a time, actively inflamed and

\* An attitude somewhat similar is given by a rare form of what I suppose to be general chronic rheumatic arthritis of the spine involving its articulations with the ribs. The spine droops and is stiff, the chest is narrow, the ribs scarcely move, the abdomen is low and broad, but there is no deformity of head or limbs.

its cavity was distended with fluid. But the inflammation soon subsided, only leaving the joint stiffer and more bent.

About this time some signs of insufficiency of the mitral valve were observed, but the patient now lived so quietly, and moved with so little speed, that this defect gave him no considerable distress.

In December, 1872, sight was partially destroyed by retinal hæmorrhage, first in one eye, then in the other,\* and at nearly the same time he began to be somewhat deaf. In the summer of 1874 he had frequent cramps in the legs, and neuralgic pains, which were described as "jumping over all the upper part of the body except the head," but change of air seemed to cure them.

In January, 1876, he began to complain of pain in his left forearm and elbow which, at first, was thought to be neuralgic. But it grew worse, and swelling appeared about the upper third of the radius and increased rapidly, so that, when I saw him in the middle of February, it seemed certain that a firm medullary or osteoid cancerous growth was forming round the radius.

Still the general health was good. Auscultation could detect mitral disease, but the appetite and digestion were unimpaired, the urine was healthy, the mind as clear, patient, and calm as ever. As letters about him at this time said "his general health has been excellent;" "he is free from pain except in the left arm; he sleeps well, enjoys himself, and does not know what a headache is."

After this time, however, together with rapid increase of the growth upon the radius, there were gradual failure of strength and emaciation, and on the 24th of March, after two days of distress with pleural effusion on the right side, he died.

\* Mr. Brudenell Carter saw him in January, 1873, and observed "the right retina sprinkled with small dots of arterial hæmorrhage, chiefly in parts remote from the centre;" and "there was no other change." The left retina was at this time healthy, but in February Dr. Clifford Allbutt found "several little plugs" in its vessels.

The body was examined five days after death, and showed no marked signs of decomposition. As it lay on a flat board its posture was remarkable, for the head was upraised to the level of the sternum, being supported by the rigid and arched spine, and the lower limbs, with the knees bent and stiff, rested on the heels and nates.

The pericranium, dura mater and all the substance of the brain appeared healthy.

The right pleural cavity contained at least a pint of pale serous fluid, with flakes and strings of inflammatory exudation. The lung was compressed, and in its pleural covering were numerous small nodular masses of pale cancerous substance. The proper pulmonary structure appeared healthy, and so did the left lung and its pleura, except that in the pleura and anterior mediastinum there were many small masses of cancer.

The heart was enlarged but thin-walled. The tricuspid and pulmonary valves and artery were healthy; the mitral valve was opaque, contracted, stiffened with atheromatous and calcareous deposits.

The aortic valves were slightly opaque but pliant, and both in them and in the first part of the aorta were numerous small patches of atheroma.

The liver and digestive canal and kidneys, examined externally, appeared healthy.

The right femur, the left tibia, the patellæ, and the upper part of the skull, were taken for separate examination, and will be separately described.

In the other bones of the skeleton, except the left radius, no signs of disease appeared externally, but I regret that they were not all more carefully examined, for I think that, at least in the clavicles and pelvis, some changes like those in the long bones of the lower limbs would have been found.

The upper third of the left radius was involved in a large ovoid mass of pale grey and white soft cancerous substance, similar to that of the nodules in the pleuræ and mediastinum, but with growths of bone extending

into it. The rest of the radius and the ulna appeared quite healthy.

Some nodules of similar cancerous substance were imbedded in the bones of the vault of the skull.

Microscopic sketches of these structures by Mr. Butlin are appended (Plate II, figs. 1-3).

The curvatures of the spine and its rigidity appeared due to shortening and hardening of its fibrous structures. The vertebræ appeared healthy; there was no appearance of overgrowth or ankylosis among them.

In no part, whether near or far from the diseased bones, was there an indication of any change of structure in skin, muscle, tendon, or fascia; but in the right hip-joint and in the left knee-joint there was some thinning and wasting of articular cartilage, such as one sees in chronic rheumatic arthritis. The other hip- and knee-joints and both ankle-joints were healthy.

In the arteries of the lower limbs there was extensive atheromatous and calcareous degeneration.

The enlargement of the skull may be estimated by comparison of the following measurements:

	Diseased skull.	Average skull.
Circumference at the level of the middle of the temporal fossa . . . . .	26½ in.	21 in.
From occipital spine to base of nasal bones . . . . .	15 in.	13½ in.
From mastoid to mastoid process . . . . .	18½ in.	15½ in.

All the sutures, at least all those of the vertex, were obliterated. The outer surface of the upper part of the skull was lowly bossed by the predominant thickening of the hinder part of the parietal bones. The thickness was in every part increased to the extent shown in these following measurements.

In a median vertical section the thickness of the frontal bone was 11-13 lines.

"	"	parietal	. 14-16	"
"	"	occipital	. 8-12	"

In a horizontal section, through the middle of the temporal

fossa, the thickness of the frontal bone was . . . . .	. 8-9	"
" temporal " . . . . .	. 6-9	"
" at their junction . . . . .	. 2	"
" of the occipital bone was . . . . .	. 10-12	"

Comparing these measurements with those of average healthy skulls, it may be said that the bones of the vault of this skull were in every part increased to about four times the normal thickness.

The whole outer surface of the skull-cap was finely porous; in the least changed parts, such as the squamous bone, perforated with innumerable apertures for blood-vessels; in the most changed, finely reticulate, as with delicate cancellous and medullary texture.

The inner surface was comparatively smooth and appeared little changed, except by the enlargement of all channels and apertures for blood-vessels, and especially by the deepening of all the grooves for the middle meningeal artery and its branches.

On the cut surface, in the median vertical section, that which might be regarded as the altered internal table of the skull was a layer, having a very unequal thickness varying from two to six lines, consisting of hard white bone, close-textured, in some parts porous or finely reticulate, in more looking compact and dense like limestone or white brick (Pl. V).

The rest of the thickness of this part of the skull, representing probably the altered diploë and outer table, was made up of bone in various degrees porous, cancellous, or cavernous, with spaces filled with soft reddish substance, a kind of medulla. Its surface was covered with a very thin layer, a mere coating of more finely porous bone.

In the horizontal section, at the level of the upper part of the squamous bone, the same altered characters were observable, but a larger proportion of the substance of the skull was finely porous or reticulate.

By the cavities in the skull-cap in which cancerous growths were lodged the structure of the bone was neither more nor less altered than in other parts.

A portion of sphenoid bone showed changes of structure very similar to those already described, but with a much more uniform and regular finely porous condition.

The bones of the face were not uncovered, but they



showed, neither to sight nor touch, any appearance of disease; not a feature was unnatural.

The conditions of all the long bones were so similar that one description may serve for the altered structure of both femora and tibiæ.\*

The periosteum was not visibly changed, not thicker or more than usually adherent.

The outer surface of the walls of the bones was irregularly and finely nodular, as with external deposits or outgrowths of bone, deeply grooved with channels for the larger periosteal blood-vessels, finely but visibly perforated in every part for transmission of the enlarged small vessels. Everything seemed to indicate a greatly increased quantity of blood in the vessels of the bone.†

The medullary structures appeared to the naked eye as little changed as the periosteum. The medullary spaces were filled with soft, yellow, ruddy, and bright crimson medulla, of apparently healthy consistence. The medullary laminæ and cancelli had a normal aspect and arrangement, and in the shafts of the long bones the medullary spaces were not encroached upon.

The compact substance of the bones was, in every part, increased in thickness. Taking, for example, the femur, the thicknesses of its walls and those of a healthy femur of about the same length and age are compared in the following tables.

	Healthy	Diseased.
	Lines.	Lines.
Thickest parts of the wall . . . . .	3—6	6—10
Articular covering of head, about . . . . .	$\frac{1}{4}$	3—10
Wall of neck, about . . . . .	$\frac{1}{4}$ —3	4—6
Wall of the trochanter major, about . . . . .	$\frac{1}{4}$ — $\frac{1}{2}$	3—5
Articular covering of the condyles, about . . . . .	$\frac{1}{4}$ — $\frac{1}{2}$	3—5
Lateral walls of the condyles . . . . .	$\frac{1}{8}$ — $\frac{1}{4}$	2 and more.

Changes in similar proportions were found in the walls

\* Their changes are shown in Pl. IV. The specimens are in the Museums of the Royal College of Surgeons and of St. Bartholomew's Hospital.

† But see p. 47 in the account of the microscopic examination.

of the tibia. In the patellæ the walls were from three to five lines thick.

The thickening of the walls of the shafts of the bones appeared due chiefly to outward expansion and some superficial outgrowth. In some places there were faint appearances of separation of parts of the outer layers of the walls, and of these becoming thick and porous, while the corresponding parts of the inner layers were less changed; but in the greater part of the walls the whole construction of the bone was altered into a hard, porous or finely reticulate substance, like very fine coral. In some places, especially in the walls of the femur, there were small, ill-defined patches of pale, dense, and hard bone looking as solid as brick.

In the compact covering of the articular ends of the long bones, and in those of the neck and great trochanter of the femur, and in the patellæ the increase of thickness was due to encroachment on the cancellous texture, as if by filling of its spaces with compact porous, new-formed bone.

Mr. Butlin was so good as to make careful microscopic examination of the diseased bones, and to give me the following report on them, together with the annexed drawings of their minute structure.

“Microscopical examination was made of sections cut from the skull and from the tibia, some of them from the recent bones, but the majority of them from portions of bone deprived of earthy salts and rendered sufficiently soft to be cut with a razor. The appearances observed were essentially the same in both bones, but most of the drawings and description were taken from the tibia, the sections of which were much clearer than those of the skull.

“The examination was conducted from a twofold point of view: first, to discover the changes which the bone had undergone; second, to discover, if possible, the nature of the process which had led to such changes.

“With a low power the number of Haversian systems

and canals in any given section was seen to be much diminished (Plate II, fig. 8; Plate III, fig. 9). The space between the Haversian canals was occupied by ordinary bone-substance, containing numerous lacunæ and canaliculi. The Haversian canals were enormously widened, many of them were confluent, and thus the appearance of a number of communicating medullary spaces was obtained, an appearance which was rendered still more striking by the presence in the canals of a large quantity of ill-developed tissue in addition to the blood-vessels (Plate II, figs. 4—6). With a high power the contents of the Haversian canals were seen to consist generally of a homogeneous or granular basis, containing cells of round or oval form about the size and having much the appearance of leucocytes. Larger nucleated cells were also present, and fibres or fibro-cells, sometimes in considerable quantity. Myeloid cells were occasionally observed, but they were not plentiful; fat also existed in many of the larger spaces, especially in the skull. The vessels were usually small compared with the channels in which they ran; indeed, they did not seem to be much larger than those of normal bone (Plate II, fig. 6). The walls of some of the canals were lined by a single layer of osteoblasts, a condition precisely similar to that observed in the normal ossification of bone in membrane. The presence of new bone was most evident in the periosteum of the tibia, external to the ordinary compact layer of the shaft (Plate II, fig. 7). This external layer was, of course, but thin, and was much softer and less developed than the cortex of the bone from which it sprung; it evidently was not nearly sufficient to account for the great increase in the diameter of the tibia. From the diminution in size of the medullary canal it was thought that a similar recent formation of bone would be found on its outskirts, but this expectation was not justified by observation.

“With a medium power the number of (Plate III, fig. 12) lamellæ surrounding the Haversian canals was

easily seen to be not larger than in normal bone, whilst the arrangement of the intervening space was most complex and totally different from that of healthy bone. The lacunæ and canaliculi throughout the sections did not strikingly differ from those of ordinary bone."

I am indebted to Dr. Russell for the following chemical analysis of portions of the diseased skull and tibia, and of a healthy tibia in comparison with them.

	Skull.	Tibia.	Normal tibia.
Inorganic constituents (Ash) . . .	60.59	61.22	63.62
Organic " . . .	39.41	38.78	36.38
Phosphoric acid ( $P_2O_5$ ) . . .	22.76	25.45	25.50
Carbonic " ( $CO_2$ ) . . .	3.59	3.95	3.59
Fat . . .	6.83	3.45	—
Moisture in the sample (dried at $115^\circ C.$ ) .	15.49	11.83	9.73
The $CO_2$ calculated as calcium carbonate ( $CaCO_3$ ) . . .	8.17	8.99	8.16
The $P_2O_5$ calculated as calcium phosphate ( $Ca_3P_2O_8$ ) . . .	49.70	55.56	55.66
Specific gravity . . .	1.895	1.889	1.886*

Cases of the disease which I have described are so rare that I believe no one has seen a sufficient number of them to enable him to distinguish this disease, either clinically or anatomically, from some which seem like it. Specimens illustrating it are commonly included under a general name of hyperostosis, osteoporosis, senile rachitis, or the like. But I hope that, if I add to the description I have just given some notes of similar cases which I have seen or found on record, the disease may be so distinguished as to deserve in pathology a separate place and name.

CASE 2.—Some ten years ago I saw a gentleman, between fifty and sixty, very active, tall, thin, and muscular, a master of hounds. For many years before his death he had curvature of the thighs and legs, exactly like that

\* Specific gravity of normal skull 1.990.

already described, and stooping of the spine. The changes of the limbs were attended with severe pains, which he used to relieve with hard rubbing, but the general health was unimpaired. In the last years of his life the upper part of his right humerus became very large, and as he was riding and suddenly raised his arm the bone broke near the shoulder. The evidence of large tumour now became clear, and I amputated the arm at the shoulder-joint. The tumour was well marked and very vascular medullary cancer investing and infiltrating the upper part of the humerus. The rest of the humerus was healthy, and the fracture, which was just below its neck, was evidently due to muscular force acting on its structures spoiled by the cancerous growth. He died a few days after the operation, but was not examined after death. The similarity of his case with that which I have described is, I think, certain.

CASE 3.—I saw, with the late Dr. Brinton, a gentleman between forty and fifty who may be still living. He was a sturdy and quite healthy man; his tibiae were curved and enlarged exactly like those in the first case and he had similar pains, but there was more thickening of periosteum and an appearance of more external formation of bone. He was treated with iodide of potassium and many other things as for periostitis, but without avail.

CASE 4.—A case is recorded by Dr. Wilks in the 'Transactions' of the Pathological Society,\* and through the kindness of Sir William Gull, whom the patient occasionally consulted, I am enabled to add some facts to those in Dr. Wilks's report, and to show photographic portraits.

A summary of Dr. Wilks's report is that the patient was sixty when he died. Signs of the disease, beginning with pains like those of rheumatism in the legs, were first observed fourteen years before his death. It was soon found that the tibiae were enlarged, and in subsequent

\* Vol. xx, p. 273, 1869.

years the cranium and nearly all the bones of the skeleton underwent similar changes. About a year before death the general health began to suffer from the thorax having become implicated in the disease. Gradually the chest became more contracted and at last quite fixed; the breathing became more difficult until at last the respiratory apparatus altogether stopped.

Sir William Gull's notes tell that the patient consulted him when fifty-six years old, and said that he first noticed enlargement in the left tibia when he was forty-five years old; that he had seven brothers well and strong, and was eldest in the family. He complained chiefly of weakness, inability to make exertion, feeling of nervousness with occasional vertigo, shortness of breath, stiffness in neck, hoarseness and feebleness of voice. His general health was good; he was not much troubled with pain anywhere; but had occasional strange sensations about the head, and much cough. His height, when a young man, was five feet three and a half inches, now four feet eleven and a half inches. The urine was normal and of normal colour. The cranium was enlarged and thickened; the clavicles much thickened, as also the long bones; the phalanges and facial bones and perhaps the lower jaw were not altered. The ribs were thick and immovable, as was also the sternum. There was general dulness over the chest on percussion. The respiration was chiefly diaphragmatic.

Less than a year before the patient's death Sir William Gull recorded that he was breathless, and had occasional attacks of mental confusion in which he remarked that he could not understand the sense of words. His voice was hoarse and feeble, and the hyoid bone seemed thickened. The head had continued to enlarge, and he maintained that he was still losing in height. The neck was fixed, and somewhat forward. All the viscera appeared normal. The urine, repeatedly examined, was always found normal, and of normal colour.

The record of the post-mortem examination by Dr. Goodhart leaves no doubt that the disease in this case

was the same as that which I have described, and it may be important that this patient also had cancerous disease. "A growth . . . corresponding to the growth described as epithelioma of the arachnoid surface of dura mater," grew from the inner surface of the dura mater, was as large as a chestnut and made a pit in the brain near the left Sylvian fissure.

The description of the changed structure of the bones, for which I may refer to the 'Pathological Transactions,' seems to me to indicate that the disease was more advanced in the direction of degeneracy than that which I have described, or that it had not been in any degree repaired.

CASE 5.—I owe to Mr. Bryant the opportunity of seeing a similar case which was under his care in Guy's Hospital, and of which Mr. Viney was so good as to give me notes.

The patient was a carpenter, sixty years old, a hard-working married man, and had seven children. When about sixteen years old he had a slight attack of gonorrhœa, but without sores, and no history of syphilis could be learned. When thirty-five years old he received an injury to his pelvis. Shortly after this he had trouble with his bladder, which became much distended; a large quantity of clotted blood was washed out. He lay in bed for this six weeks, and at the end of three months was able to go to work again.

For the last five years he had been troubled with gout in his left great toe. His father suffered from this. The attacks had been short; a few days' rest always sufficed for recovery.

About three years before admission he first felt pains of a shooting description about the tendons of the popliteal space, whenever he straightened his legs. At this time also he first noticed a swelling of the legs, which began at the ankles. These symptoms, without his taking any special notice of them, continued for about a year.

In the last year and a half the tibiæ had become much swollen and curved forwards, and on account of the pain

he had in them from standing he had been obliged to give up his regular work. Until admission he did not notice anything wrong with his other bones, but he had lost about half an inch in height.

The tibiæ presented a marked curve forwards. The anterior border of each was rounded to a very marked degree, so that it could not be felt at all distinctly. The right tibia was slightly larger than the left. The inner surface of each measured about four inches at its widest part. The veins above the ankle were in a varicose condition.

The fibulæ were very much enlarged; the femora enlarged in their shafts and bowed outwards. The great trochanter was drawn up to the level of a vertical line drawn from the anterior superior spinous process of the ilium to the horizontal line of the body, instead of being about two and a half inches below this line. The patellæ were little larger than natural.

The bones of the upper extremity were enlarged, but not to so marked a degree as those of the lower. The enlargement was most marked in the humeri and the left was thicker than the right. He could not straighten his arms, probably owing to the enlargement of the olecranon. In the clavicles the natural curves were very much increased and the bones thickened, the left more so than the right. In the scapulæ the spines and acromion processes were very much enlarged.

The chest was slightly flattened from side to side, but moved fairly whilst breathing. The ribs on the right side were slightly larger than those on the left.

There was a general curve backwards from the cervical to the dorsal vertebræ, so that the patient's usual position in bed was with his head bent forwards, and his legs in a semi-extended position.

The bones of the hands and feet did not seem to have shared in the general thickening.

There seemed to be a slight thickening about the external protuberance of the occipital bone, but there was no other evidence of the cranial bones being involved.



The patient had cold perspirations over his legs in the evening. His urine had a specific gravity of 1014, was strongly acid, contained a little albumen but no excess of phosphates.

[Six months later Mr. Bryant told me that this patient's bones were still enlarging, and that there were evidences of enlargement of the skull.]

I have looked for records of cases similar to these in nearly every work that seemed likely to contain them, but in vain. I have found only three cases, and the first two of these are doubtful.

Saucerotte\* relates the case of a man who died at forty and in whom all the bones, those of the head, face, orbits, ribs, vertebræ and limbs had begun to enlarge about seven years before death. He increased in weight from 119 livres to 168, wholly from increase of bones; he had rheumatic pains; for a time sleepiness, oppression at the chest, and very small pulse; but these passed-by and he died with some acute illness. No examination was made.

Rulliert† tells of a man, aged seventy-eight, who died in the Hôtel Dieu of empyema. He had previously been in good health, and nothing had indicated any derangement of cerebral function. The skull was very large, osteoporotic, and heavy, and, except the lower jaw, all the bones of the face were healthy. The ribs were thicker and larger than usual; the sternum narrow and very thick; the pelvic bones changed like those of the skull. The clavicles were thick, curved, and solid. The other bones were healthy.

Wrany‡ has fully described the condition of the bones in a case of spongy hyperostosis of the skull, pelvis, and left femur, taken from a woman fifty years old, of whom, however, nothing is told but that she died of pyæmia, and

\* 'Mélanges de Chirurgie,' Paris, 1801.

† 'Bulletin de l'École de Médecine de Paris,' t. ii, p. 94, 1812.

‡ 'Prager Vierteljahrschrift,' 1867, B. i, p. 79.

that she had "spongy hyperostosis of the skull with atrophy of the facial skeleton, spongy hyperostosis of the vertebral column, pelvis, and left femur, with elongation of the latter bone; kyphoscoliosis of the upper dorsal part of the spine; pelvic abscess; emphysema and œdema of both lungs, abscess of the left; marasmus."

I cannot doubt that this disease was the same as I have here described, and the paper is valuable, both for the many signs indicated in it that the bones softened and yielded to pressure in the early part of the disease, and for the careful comparison of the distortion of the pelvis with the dissimilar distortions in rickets and mollities ossium. The spine was very curved; the chest small and too arched; the whole trunk very short.

From these cases which, though few, are well marked and in some chief points uniform, as well as from a recollection of two more of which I have no notes, I think we may believe that we have to do with a disease of bones of which the following are the most frequent characters:— It begins in middle age or later, is very slow in progress, may continue for many years without influence on the general health, and may give no other trouble than those which are due to the changes of shape, size, and direction of the diseased bones. Even when the skull is hugely thickened, and all its bones exceedingly altered in structure, the mind remains unaffected.

The disease affects most frequently the long bones of the lower extremities and the skull, and is usually symmetrical. The bones enlarge and soften, and those bearing weight yield and become unnaturally curved and misshapen. The spine, whether by yielding to the weight of the overgrown skull, or by change in its own structures, may sink and seem to shorten with greatly increased dorsal and lumbar curves; the pelvis may become wide; the necks of the femora may become nearly horizontal, but the limbs, however misshapen, remain strong and fit to support the trunk.

In its earlier periods, and sometimes through all its

course, the disease is attended with pains in the affected bones, pains widely various in severity and variously described as rheumatic, gouty, or neuralgic, not especially nocturnal or periodical. It is not attended with fever. No characteristic conditions of urine or fæces have been found in it. It is not associated with syphilis\* or any other known constitutional disease, unless it be cancer.

In three out of the five well-marked cases that I have seen or read-of cancer appeared late in life ; a remarkable proportion, possibly not more than might have occurred in accidental coincidences, yet suggesting careful inquiry.†

The bones examined after death show the consequences of an inflammation affecting, in the skull the whole thickness, in the long bones chiefly the compact structure, of their walls, and not only the walls of their shafts but, in a very characteristic manner, those of their articular surfaces.

The changes of structure produced in the earliest periods of the disease have not yet been observed, but it may certainly be believed that they are inflammatory, for the softening is associated with enlargement and with excessive production of imperfectly developed structures, and with increased blood-supply. Whether inflammation in any degree continues to the last, or whether, after many years of progress, any reparative changes ensue, after the manner of a so-called consecutive hardening, is uncertain.

The inflammatory nature of the disease is evident also in the changes of minute structure in the affected bones.‡

\* There has not only been no history of syphilis in any of the cases, but no known syphilitic changes have been observed in any patient.

† See, also, Sandifort, quoted at p. 61 ; Museum of St. Bartholomew's, ser. i, 111 and 112, sections of a femur, large, curved, porous, with a tumour growing around its shaft ; and 49, a hyperostotic skull from a man who died with cancerous disease of the eyeball, heart, and other organs ; and Museum of Guy's Hospital, specimens of symmetrical osteoid cancer of the ilia, with cancer of the spine and cranium, associated with hypertrophy of the cranium. Dr. Goodhart was so good as to give me a report of this case.

‡ And this is also the opinion of Wrány, l. c.

On these Mr. Butlin writes, "With regard to the nature of the process by which these changes were accomplished, there are probably only three things which could produce so great an increase in the size of a bone, namely, new growth (tumour), hypertrophy, and chronic inflammation.

"The first of these may be at once set aside as out of the question.

"Nor is the second much more probable than the first, for the process is evidently no mere hypertrophy. The whole microscopical architecture of the bone has been altered; the structure appears to have been almost entirely removed and laid down afresh on a different plan and in a larger mould.

"Of the three causes chronic inflammation alone remains, and upon examination one or two facts will be found to bear strongly upon the theory of this being essentially an inflammatory disease. Not only the absorption of the old structure which has taken place, but also the manner of this absorption, point to its inflammatory nature. Traces of this are not, of course, always discernible, as the process is almost everywhere far advanced. But still careful observation not uncommonly discovers that the sides of the widened canals, instead of being smooth and even (Plate III, fig. 10), are eaten out in a series of curves or concavities with the production of what are called Howship's lacunæ, so characteristic of inflammation. The tissue contained in the canals, too, almost precisely resembles the tissue found in the spaces of inflamed bones, only differing from it in being generally more fibrillar and less rich in cells, a fact easily to be accounted for by the very long duration of the disease and the general tendency towards organisation which was displayed throughout. The apparent cessation of the process of absorption and the gradual process of repair may be regarded as still further leading towards the same conclusion.

"Further than this the microscopical observations do not extend."

The chemical analysis by Dr. Russell may be regarded as confirming this conclusion. It shows, at least, that there is no such change of composition in the bone as would be expected in any merely degenerative softening.

Holding, then, the disease to be an inflammation of bones, I would suggest that, for brief reference, and for the present, it may be called, after its most striking character, *Osteitis deformans*. A better name may be given when more is known of it.

It remains that I should point out the distinctions between this disease and the several forms of hyperostosis, osteoporosis, and other diseases among which it has been confused.\*

1. Among cases of hyperostosis are included those of simple overgrowth or hypertrophy of bones in adaptation to increase or change of office. The distinction of these from any form of disease is plain enough; they show a mere increase of natural structure.†

2. Scarcely different from these and as easily distinguished are the hyperostoses, best seen in the skull, in which the bones have more than normal thickness, hardness, and weight, and marks of greater vascularity, yet preserve a just relation of their several parts and a scarcely changed structure. They probably illustrate the effects of simple inflammation of bone recovered from.‡

3. A group of hyperostoses consists of those cases in which bones are enlarged in consequence of an increased supply of blood or lymph. Such a case is that recorded by Dr. Day§ in which the bones of a boy's limb with obstructed lymphatics are much longer than those of the sound limb;|| and such are all those in which bones near

\* Many of the statements here made are derived from the examinations of the collections of diseased bones in the College of Surgeons and St. Bartholomew's Hospital, which I made while writing the catalogues of their pathological museums.

† Mus. Coll. Surg., 379, 380, 2838, 2839, 2842, 2843, &c.

‡ Mus. Coll. Surg., 2840, 2841.

§ 'Transactions of the Clinical Society,' vol. ii, p. 104, 1869.

|| Broca, 'Des Anévrysmes,' 8vo, p. 76, 1856, gives a case of femoral arterio-venous aneurism attended with considerable elongation of the limb.

inflamed joints, or with partial necrosis, or in limbs long hyperæmic, from whatever cause, grow in length and circumference till they considerably surpass the bones of the healthy limb.\* These are easily distinguished. They have not signs of disease proper to themselves; they occur in the young alone; they may present a healthy texture, or one only slightly changed as by partaking of the adjacent inflammatory process; and with the exception of the tibia they do not become deformed. The tibia, when it lengthens more than the fibula, is almost compelled to curvature by the fixed unyielding attachment of its ends;† and the curve is usually similar in shape and direction to the curve of the tibia in the osteitis deformans. But there is no other likeness between the two conditions.

4. A very large number of cases of hyperostosis are consequences of inflammations of bone; some of simple inflammation, others of scrofulous, syphilitic, or gouty inflammation. It is not necessary here to distinguish these from each other,‡ but there are sufficient signs for the distinction of all from the osteitis deformans.

It is clear that the summary which I have given of the clinical characters of this osteitis would not tally with that of any case of simple osteitis, such as might ensue in a healthy person after injury, or in the neighbourhood of a sequestrum; and the clinical difference is as complete between it and any case that could justly be regarded as strumous or syphilitic or gouty osteitis.

The anatomical differences are as well marked: chiefly

\* I believe these were first described by Mr. Stanley, 'On Diseases of Bones,' p. 20, *et seq.*, and myself, 'Lectures on Surgical Pathology,' p. 64, ed. 3, and in the catalogues already referred to. Langenbeck has published a very interesting paper on them in the 'Berliner Klin. Wochenschrift,' 1869, No. 26. Cases are also cited from Weinlechner, Schott, and Bergmann, in Virchow and Hirsch's 'Jahresbericht für 1869.'

† Such curved tibiæ are in the museum of St. Bartholomew's, Nos. A. 3, A. 46.

‡ An attempt to do so is made in the pathological catalogue of the College of Surgeons.

in the facts that in these inflammations the bones do not become curved\* (unless in the case of the tibia already explained); that they commonly display much more considerable external periosteal outgrowths or deposits, as if from a greater participation of the periosteum in the inflammatory process; that the rarefied or, it may be, porous structure of the swollen shafts of bones usually shows appearances of separation and expansion of the component layers; that the medullary canals are commonly invaded by the thickening walls, or are as much changed as the walls themselves; that the whole length of a bone-shaft is very rarely affected; and that the thin articular layers of bones are, I believe, never thickened as they are in the osteitis deformans.†

It may be added that it is very improbable that any form or degree of scrofula or syphilis or gout should exist in bones or any other textures for ten or more years without affecting other parts and without impairing the general health. The retention of good general health during many years of localised disease is, indeed, one of the most striking characters of the osteitis deformans. The only parallel known to me is in the rheumatoid or chronic rheumatic arthritis, and the likeness between the two in this respect may suggest that they are nearly related; yet they are not found concurrent. In the case that I have related the amount of chronic rheumatic arthritis was trivial, and (which is more important) in all the records and specimens of the arthritis which I have seen I have not found an instance in which there were any of the morbid changes characteristic of the osteitis.‡

5. There are, I think, only two other diseases, namely,

\* The absence of curving in bones around sequestra is remarkable, for they are long and often acutely inflamed, and those of the lower limbs are commonly used and bear weight.

† Among the specimens in which these changes may be studied are, in the College Museum, Nos. 3085, 3089, 598, 3090, 3091; in the museum at St. Bartholomew's, A. 1, and ser. i, 56, 132, 138, 196—198.

‡ There is not even any mention of them in Mr. R. Adams's elaborate 'Treatise on Rheumatic Gout,' 1873, 8vo and folio.

rachitis and osteomalacia, from which it can be necessary to discriminate the osteitis deformans, and the differences between them are very wide. They have scarcely a feature in common except that in all of them the bones bearing weight become curved or misshapen, and the spine is usually deformed, and the skull may become very thick and porous. But in rachitis the bones are too short, not too long; too small, not too large; and their curvatures are quite unlike those of the osteitis. And in the osteomalacia the walls of the bones become exceedingly thin, wasting with an acute atrophy; and when they yield it is not with regular curving but with angular bending or breaking. By these and many other differences, as well clinical as anatomical, the diagnosis of the osteitis from rachitis and osteomalacia is sufficiently clear. With rachitis it may be judged to have no affinity whatever; with osteomalacia only so much as may exist between a chronic inflammation and an acute atrophy of any part. Yet by one character which all these three diseases have or may have in common, namely, the osteoporosis of the skull, they are constantly confounded in museums, if not in practice, with each other, and with diseases different from them all.

The study of the osteitis deformans led me to learn what I could of the various recorded descriptions of large, thick, and porous skulls often found in museums. Nearly every large museum contains one or more specimens of such skulls whole or in fragments. They are all big, thick, porous, or spongy, with obliterated sutures and wide apertures and grooves for blood-vessels. Very few of these specimens have any life-histories; they are all, in many respects, alike and usually are all named alike. Many of them it may be impossible to name or classify without much better knowledge of them than may now be had, but I believe that among them are the results of several different diseases; and it may save some trouble to future students if I refer to some of the specimens and records which have led me to this belief.



1. Some are examples of the osteitis deformans which I have described.\*

2. Some are derived from cases of osteomalacia. Mr. Durham† has written on these, and Mr. Solly's‡ well-known paper gives a good instance of them. In general, I think that these may be distinguished, at least in the recent state, by their softness and lightness; the abundance of soft medulla contained in them, and the comparative brittleness of the bones when dry.

3. Some are from rachitis; they are, unless after recovery and repair, very light, almost friable, and on their surface not porous, but like fine cloth or felt.§ Like these are the skulls of some lions and monkeys which have died young, in confinement, of what is considered rickets. A collection of these skulls and other similarly diseased bones in the college museum|| deserves careful study, especially because of their likeness to the cases included in the next group.¶

4. These are the results of a disease of early life, sometimes even of childhood, in which all the bones of the face as well as those of the cranium are affected, and, it is said, the bones of the limbs. All the affected bones, facial as well as cranial (and herein is a clear ground of diagnosis), become hugely thickened, porous, or reticulate.

\* To those already referred to these, I think, may be added: Sandifort, 'Museum Anat. Acad.,' Lugd.-Bat., fol., 1835, vol. i, p. 142, vol. ii, tab. xiii, Skull of a man forty-three years old, with a "fungus" over the left orbit (? a cancerous growth). Other similar skulls are here referred to. Similar specimens are, probably, Nos. 2840 and 2858A in the College Museum; and, more uncertainly, 2841 and 2858, which, perhaps, belong rather to the fifth group.

† 'Guy's Hospital Reports,' ser. iii, vol. x, 1864.

‡ 'Med. Chir. Trans.,' vol. xxvii, p. 435, Mus. Coll. Surg., 395.

§ See Mus. Coll. Surg., 390—394, 2844, and 2857. I believe that Huschke, 'Ueber Craniosclerosis,' 1858, quoted by Virchow, contains facts on the rachitic osteoporoses, but I have not been able to refer to it.

|| Nos. 386—388, 2854—2856, 2855A, &c.

¶ Although bones such as these are not described by Paul Gervais, yet his paper quoted below should be studied on all that relates to hyperostosis in animals.

The whole skull is very large, clumsy, and featureless. Commonly the cranial cavity is diminished. The orbital and nasal cavities are contracted, the antra are often filled, by the ingrowth of their several walls; the apertures for nerves are narrowed or obliterated.\*

Of these cases, which are among those named by Virchow† *Leontiasis ossea*, the best are related by Ilg‡ and Jadelot.§ Their descriptions are very scanty, yet they give sufficient facts to distinguish the disease by their account of the cerebral symptoms associated with it. In Ilg's case, for example, the patient who died at twenty-seven, after seventeen years' disease, had amaurosis, epilepsy, severe general headache, delirium, convulsive attacks, and at last total deafness, witlessness, difficulty of swallowing, and loss of smell.

5. Some cases, perhaps not different from these, though they have occurred in later life, are those by Schützenberger,|| Otto,¶ and Wrány.\*\*

6. And, lastly, there are cases not so much of thickening

\* Among the casts in the museum at St. Bartholomew's, No. 10, is that of a skull affected with this disease, and in ser. i, 36 are fragments of a bone, which, I think, may be referred to it.

† 'Die krankhaften Geschwülste,' B. 11, 1864-5. I need not say that this contains a very complete account of all forms of overgrowth of bone.

‡ 'Einige Anatomische Beobachtungen,' 4to, Prag, 1821.

§ Quoted by Ilg from Meckel. The best of many accounts of this specimen is given by Paul Gervais, "De l'hyperostose chez l'homme et chez les animaux," in the 'Journal de Zoologie,' t. iv, 1875. He has carefully re-examined the skull and face and described them.

|| 'Gazette Médicale de Strasbourg,' and in Canstatt's 'Jahresbericht für 1856,' B. iii, 34, with references to cases by Breschet and Nélaton.

¶ Otto, 'Neue seltene Beobachtungen,' 4to, 1824, p. 2. Both head and face are affected; the bones are described as, after softening, very hard, dense, and almost ivory-like. Six hyperostotic skulls are mentioned in his 'Neues Verzeichniss der Anat. Sammlung zu Breslau,' 1841.

\*\* Wrány, "Hyperostosis maxillarum," in 'Prager Vierteljahrschrift,' 1867, B. 1, similar affections of the facial and cranial bones, with cerebral symptoms. Doubtful cases by Ribelt are quoted by Ilg, l. c.; Malpighi, 'Opera Posthuma,' 4to, Amstel., 1700, p. 68; Kilian, 'Anat. Unters. über den neunten Hirnnervenpaar,' Pesth, 4to, 1822, p. 133; Quekett, reported by Hewett, 'Medical Times and Gazette,' Sept. 8th, 1855, p. 229.

of the cranial and facial bones as of enormous bossed and nodular hard bony outgrowths overspreading them or projecting from them. The leading case among these is that published in the 'Transactions' of the Pathological Society by Dr. Murchison,\* with a report on the specimens by Mr. De Morgan and Mr. Hulke.† The disease in which the facial more than the cranial bones are affected is clearly distinct from any of the foregoing, or if it be in any way connected with them, especially with those of the fifth group, may be regarded as transitional from them to the exostoses, especially the massive tuberos and bossed ivory exostoses, which grow on or among the bones of the face and skull. The same approach to the character of hard exostoses is shown in the disease of the fibula in Dr. Murchison's case, a section of which, from the museum of the Middlesex Hospital, is now before the Society.

\* Vol. xvii, 1866, p. 243.

† Similar cases are illustrated by Forcade, quoted in Virchow's 'Die krankhaften Geschwülste,' B. 2, p. 22; Weber, from a specimen in the Dupuytren Museum, in v. Pitha and Billroth's 'Handbuch,' B. 3, Abth. 1, Lief ii, p. 257; Howship, 'Practical Observations in Surgery and Morbid Anatomy,' 1816, p. 26; Adams in 'Trans. of the Pathological Society,' vol. xxii, p. 204, 1871; Lysthay, in Canstatt's 'Jahresbericht für 1858,' Mus. Coll. Surg. Eng., 3093. Virchow has a full account of nearly all these cases, and of the analogies of the disease with elephantiasis of soft parts.

## DESCRIPTION OF PLATES I TO V.

Chronic inflammation of bones (Osteitis deformans).

PLATE I. See pp. 39-40.

Figs. 1—3. From photographs of the patient (Case 1) taken six months before death.

Fig. 4. From photographs of the same patient's cap worn in 1844, and hat worn in 1876.

PLATE II. See pp. 42-3 and 46.

Figs. 1, 2. From tumour of forearm. Fig. 1. Oc. 3, obj. 4.  $\times$  about 62. Fig. 2. Oc. 3, obj. 7. Tube drawn out.  $\times$  260.

Fig. 3. From secondary tumour of pleura. Oc. 3, obj. 7, t. dr. o.  $\times$  260. See p. 42-3.

Figs. 4, 5. To show tissue in widened canals of tibia (4) and skull (5). Oc. 3, obj. 7, t. d. o.  $\times$  about 260.

Fig. 6. Trabecula of bone (tibia) lined by osteoblasts.  $\times$  about 260.

Figs. 7, 8. From transverse section of tibia. (A. i. in.) Fig. 7 shows new bone growing in periosteum. Fig. 8. Taken from immediately beneath the periosteum.

PLATE III. See pp. 46-7.

Fig. 9. From perpendicular section of skull. (A. i. in.)

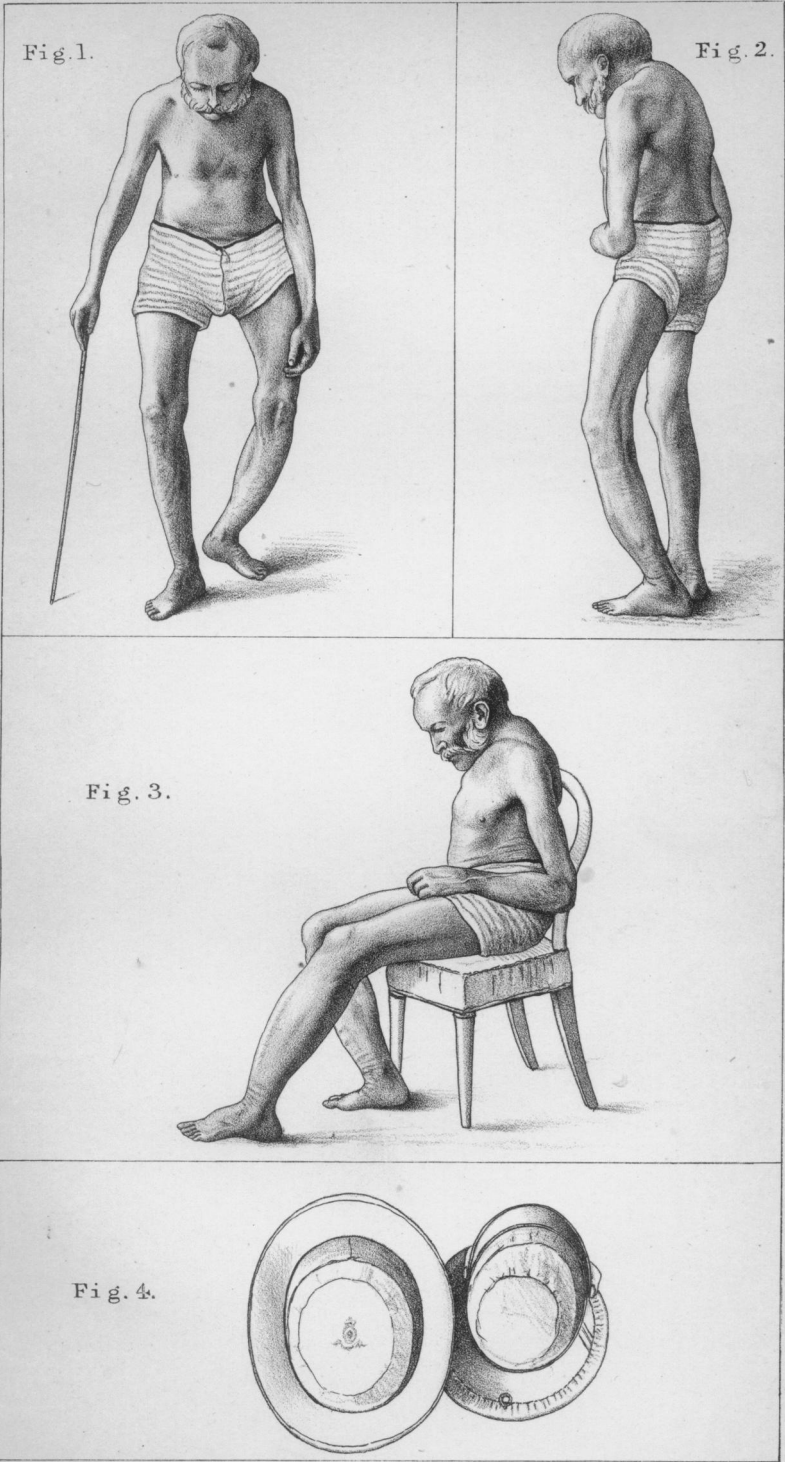
Fig. 10. From section of tibia, to show eaten-out border of widened Haversian canal. Oc. 3, obj. 7.  $\times$  200.

Figs. 11, 12. From transverse section of tibia. Fig. 11. At some distance from surface. Fig. 12. From a little way beneath the periosteum. Oc. 3, obj. 4, t. dr. o.  $\times$  87.

Fig. 13. Transverse section of normal tibia. Oc. 3, obj. 4, t. dr. o.  $\times$  about 87.

PLATE IV.—Upper and lower ends of femur. See p. 45. (College of Surgeons Museum, No. 395B. Half diameter.)

PLATE V.—Cranium. See p. 44. (College of Surgeons Museum, No. 395A. Real size.)



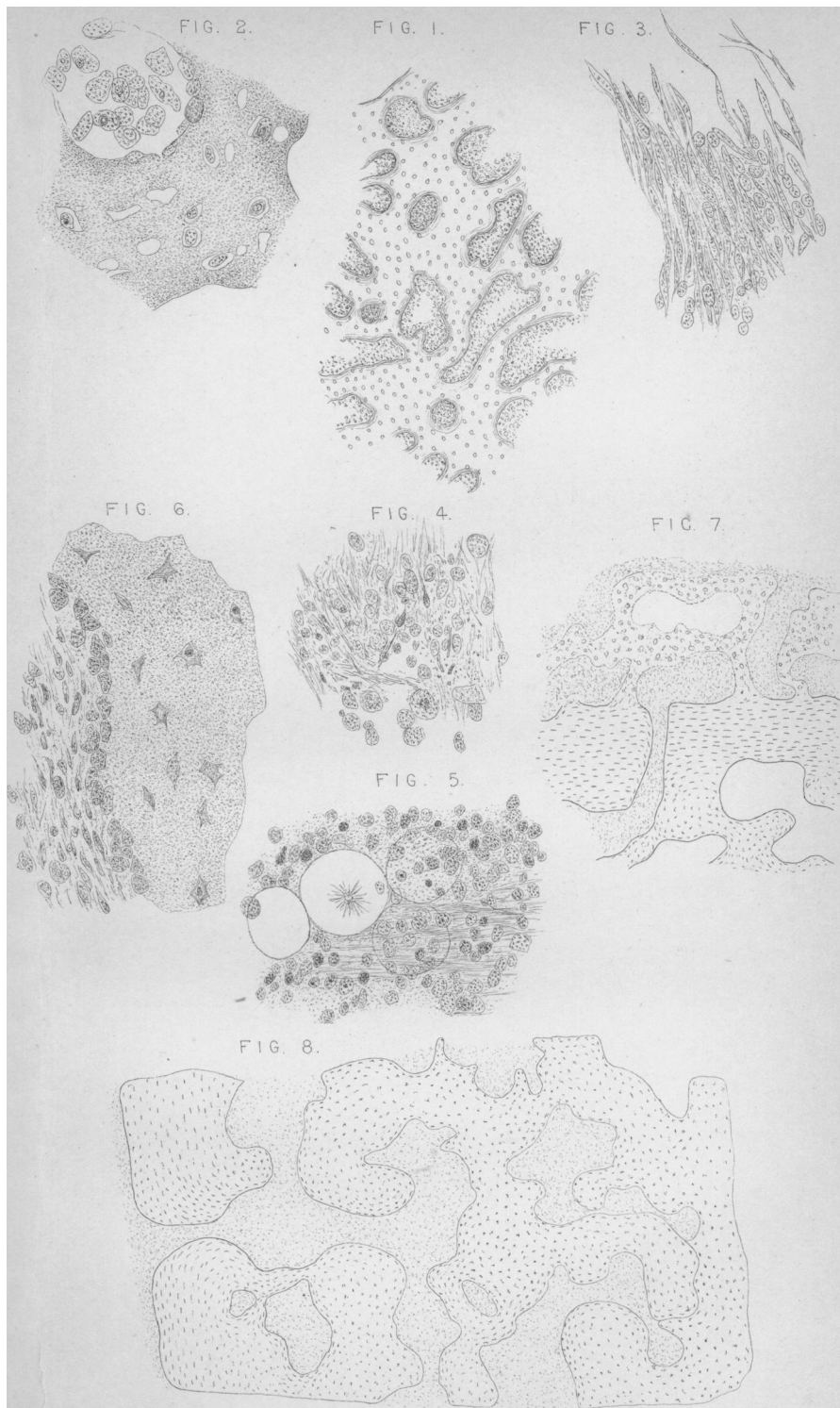


FIG. 9.

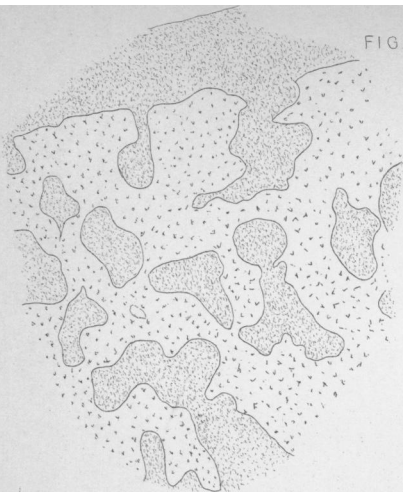


FIG. 10.

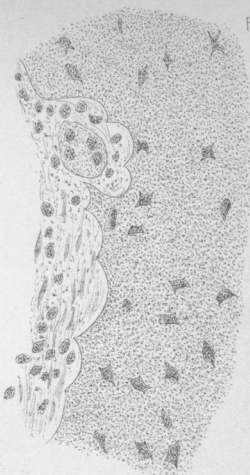


FIG. 11.

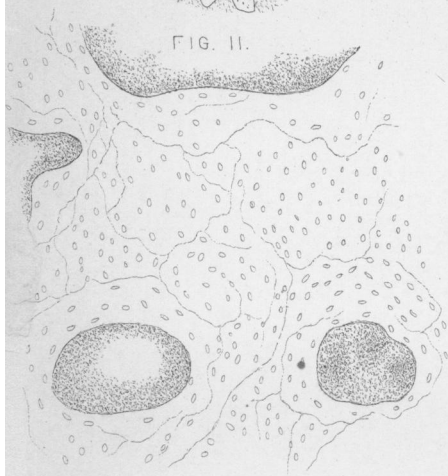


FIG. 12.

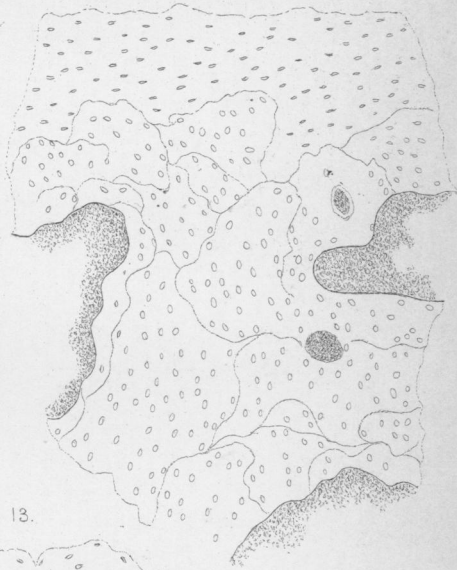
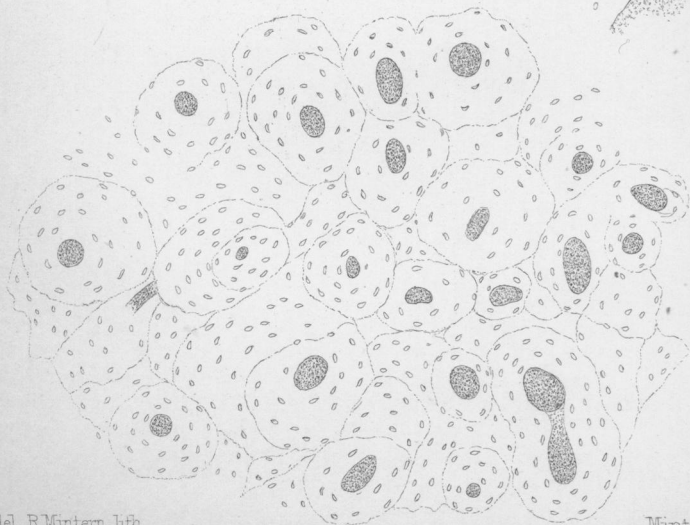


FIG. 13.





*Curve of the Femur.*



